3. Dar O, Riley J, Chapman C, et al. A randomized trial of home telemonitoring in a typical elderly heart failure population in North West London: results of the Home-HF study. Eur J Heart Fail 2009;11:319–25.

Not Just Coronary Arteritis, Kawasaki Disease Is a Myocarditis, Too

There is a great deal of insight in the points of view addressed recently by Gordon et al. (1) and by Gersony (2). One could not but notice an unbalanced aspect in the commentary article, however. This has nothing to do with the respected opinion of Dr. Gersony; on the contrary, it has to do with the general misconception connecting Kawasaki disease (KD) to merely 1 fact: the related coronary artery complications. The original editorial article by Gordon et al. (1) is intended to convey a message to the Adult Cardiology Society urging for an educated awareness of the perceived impact of the KD-related cardiovascular disturbances beyond childhood. Whether some aspects of the disease gained the deserved emphasis in the original review article or not, the rebuttal commentary was completely distracted from essential realities associated with KD. Although Gordon et al. (1) identified the often-missed importance of the consequences of KD on the myocardium, Gersony (2) limited his discussion to the sole coronary artery complications of the disease. That is exactly the problem. In my mind, it is important to put a brake on the problematic coronary aneurysms, be it only for a minute. The hidden face of the moon in this disease, the myocarditis, must not be underestimated (3) This myocarditis is evidenced by serial myocardial biopsy studies from patients without coronary aneurysms (4,5). It is also suggested by echocardiography studies (6) and by biochemical markers reflecting the myocardial response to the inflammatory process upon the onset of the disease (7). Four decades since the initial recognition of KD as a separate entity from resembling ailments have not permitted researchers to uncover its etiology. The inflammatory involvement of the myocardium and its long-term consequences deserve a serious look and a methodological follow-up.

The incidence of KD is on the rise, not only because of the modified diagnostic paradigm, which encourages the diagnosis of cases with incomplete clinical criteria (8,9), but also because of the recent awareness of the diagnosis, not only in North America but also in the most populous countries of the globe as well (10,11). It is appropriate and wise to inform KD patients that there are insufficient data to adequately calculate their cardiovascular prognosis, with the exception of the minority who sustained a severe coronary artery injury. Should we wish to care for the remaining 99% of KD patients, the myocarditis trail—not just that of the coronary arteries—needs to be followed. And as Gersony (2) correctly concludes, the American Heart Association guidelines are simply guidelines, subject to continuous updates.

Now, back to the injured coronary arteries, resolved or unresolved. If making patients aware of their antecedent KD diagnosis as a potential cardiovascular risk factor deeply affects their psyche, then physicians must also refrain from discussing obesity, for example, when counseling primary or secondary prevention of cardiovascular diseases.

***Nagib Dahdah, MD**

*Division of Pediatric Cardiology
Sixth Floor, Bloc 9
Sainte-Justine Hospital
3175 Cote Sainte-Catherine
Montreal, Quebec H3T 1C5
Canada
E-mail: nagib.dahdah.hsj@ssss.gouv.qc.ca

doi:10.1016/j.jacc.2009.11.067

REFERENCES

1. Gordon JB, Kahn AM, Burns JC. When children with Kawasaki disease grow up: myocardial and vascular complications in adulthood. J Am Coll Cardiol 2009;54:1911–20.
2. Gersony WM. The adult after Kawasaki disease: the risks of late coronary events. J Am Coll Cardiol 2009;54:1921–3.
3. Takahashi M. Myocarditis in Kawasaki syndrome: a minor villain? Circulation 1989;79:1398–400.
4. Yutani C, Go S, Kamiya T, et al. Cardiac biopsy of Kawasaki disease. Arch Pathol Lab Med 1981;105:470–3.
5. Fujiwara H, Hamashima Y. Pathology of the heart in Kawasaki disease. Pediatrics 1978;61:100–7.
6. Yu JJ, Kwak BO, Jeon YH, et al. Elevation of the index of left ventricular mass during the acute and subacute phase of Kawasaki disease, and its association with indexes of diastolic function. Cardiol Young 2009;19:64–9.
7. Dahdah N, Siles A, Fournier A, et al. Natriuretic peptide as an adjunctive diagnostic test in the acute phase of Kawasaki disease. Pediatr Cardiol 2009;30:810–7.
8. Sonobe T, Kiyosawa N, Tsuchia K, et al. Prevalence of coronary artery abnormality in incomplete Kawasaki disease. Pediatr Int 2007;49:421–42.
9. Newburger JW, Takahashi M, Gerber MA, et al. Diagnosis, treatment, and long-term management of Kawasaki disease. A statement for health professionals from the Committee on Rheumatic Fever, Endocarditis and Kawasaki Disease, Council on Cardiovascular Disease in the Young, American Heart Association. Endorsed by the American Academy of Pediatrics. Circulation 2004;110:2747–71.
10. Singh S, Kawasaki T. Kwasaki disease—an Indian perspective. Indian Pediatr 2009;46:563–71.
11. Li XH, Li XJ, Li H, Xu M, Zhou M. Epidemiological survey of Kawasaki disease in Sichuan Province of China. J Trop Pediatr 2008;54:133–6.

Reply

I thank Dr. Dahdah for his interest in my paper (1). In his letter to the editor, Dr. Dahdah raises concerns about the long-term effects of myocarditis on adults who had Kawasaki disease (KD) as a child, and that my commentary, which pertained to the risk of late coronary artery events, did not address this potential issue. As he indicates, a myocardial inflammatory process in the acute phase of KD has been well documented, even when coronary involvement may have been minimal or even absent. However, Dr. Dahdah's assertion that late manifestations of acute myocarditis are likely to be a serious threat to the adult who had KD is not evidence based. The references accompanying his letter do not describe a single case of an adult with late myocarditis or nonischemic cardiomyopathy, and there have been hundreds of thousands of patients who have had KD. Furthermore, the biopsy studies carried out in 1978 and 1981 were obtained from patients in the acute and subacute phases of the disease. By no means can

they be interpreted as sufficient evidence for physicians to raise realistic concerns about myocarditis as a significant potential problem in adults who had KD in childhood. I believe that Dr. Dahdah is in a tiny minority when he suggests that the review by Gordon et al. and, particularly, my commentary are "completely distracted from the central realities associated with KD."

Similar to the discussion concerning early transient coronary artery involvement, in >40 years of observation, the early myocardial inflammation occurring in some cases of KD has not been shown to progress to chronic myocardial disease. Of course, future clinical observations will be considered, but at this time there is no evidence to suggest that former KD patients should be instilled with the fear of late myocarditis. As in all of medicine, physician-patient communication is essential. As stated in the commentary: "The [KD] guidelines should be reviewed individually, and interpreted on a 1-to-1 basis, so that each post-KD individual will have a clear perspective as to what is actually known about late risk in his/her circumstances." Equating this statement with physicians' refraining to discuss the effects of obesity when counseling patients is not credible.

***Welton M. Gersony, MD**

*Columbia University Medical Center
3959 Broadway, 2-North
New York, New York 10032-1537
E-mail: wmg1@columbia.edu

doi:10.1016/j.jacc.2009.12.029

REFERENCE

1. Gersony WM. The adult after Kawasaki disease: the risks of late coronary events. J Am Coll Cardiol 2009;54:1921–3.

Reply

We thank Drs. Gersony and Dahdah for their thoughtful comments regarding our paper (1) on Kawasaki disease (KD) in the adult patient. There are several issues worthy of further discussion. In his perspective, Dr. Gersony agrees that the small minority of patients with giant aneurysms are at high risk for future cardiac events and deserve long-term follow-up as recommended by the American Heart Association guidelines. We suggested that "on the basis of the accumulating evidence, it is likely that patients with known aneurysms during the acute phase of KD will have some cardiovascular morbidity as young adults." Dr. Gersony responded that this is "possibly true for patients with large aneurysms but not evidence based for patients with small aneurysms." Unfortunately, the current published literature on this issue is incomplete, and the long-term outcomes for the patients in question are not really known. The majority of adults with KD will do well, but patients discharged by pediatric cardiologists with aneurysms that have "normalized" by transthoracic echocardiography have presented in our emergency room with heart failure related to severe left ventricular dysfunction. Perhaps, as Dr. Dahdah suggests, "the myocarditis should not be underestimated." Indeed, the myocardial fibrosis resulting from myocarditis associated with acute KD may turn out to be as problematic as the ischemic events related to the coronary artery disease. At this time, we simply do not have the answers to many key questions. We look forward to adequately powered, prospective, longitudinal cohort studies that will establish the numerator and denominator for the cardiovascular risk equation after KD. These studies must track not only coronary artery sequelae but also myocardial systolic and diastolic function, myocardial perfusion, valvular function, aortic root dimensions, and endothelial cell function.

We appreciate Dr. Gersony's thoughtful perspective as a senior pediatric cardiologist and agree that the majority of patients with KD fully recover from their acute vascular and myocardial insult suffered in childhood. We respectfully disagree, however, with his opinion that only patients with giant aneurysms need to be followed up longitudinally. Does the vascular injury in childhood make coronary arteries more prone to atherosclerosis and ischemic complications if the adult smokes or has diabetes? Once again, Dr. Gersony correctly observes that "there is no direct evidence" that this is the case. The absence of systematically collected data does not guarantee the absence of important problems. As adult cardiologists, we have seen young adults post-KD in childhood present with severe coronary artery disease in modest aneurysms as well as in vessels without previous aneurysms. The natural history of the vascular injury caused by KD in childhood has not yet been defined. Dr. Gersony states that it is important to avoid creating "patients" among persons who have not been shown to be at increased late risk for a cardiac event. It has been our experience that longitudinal follow-up of this patient group provides reassurance to patients and to their families rather than creating anxiety or concern.

Few adult cardiologists have managed patients with complications of KD. Failure to recognize these patients is a major problem. Our review was written with the intention of raising awareness about this remarkable disease. The eyes cannot see what the mind does not know.

***John B. Gordon, MD**
Andrew M. Kahn, MD, PhD
Jane C. Burns, MD

*San Diego Cardiac Center
Sharp Memorial Hospital
3131 Berger Street, Suite 200
San Diego, California 92123
E-mail: AdultKD@gmail.com

doi:10.1016/j.jacc.2010.01.015

REFERENCE

1. Gordon JB, Kahn AM, Burns JC. When children with Kawasaki disease grow up: myocardial and vascular complications in adulthood. J Am Coll Cardiol 2009;54:1911–20.